J Ped Surg Case Reports 3 (2015) 419–422

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

Lymphangioma involving the urogenital system in childhood

Michael Yap [a,*], Robert Newbury [b], Denise Malicki [b], George Chiang [a]

[a] Division of Pediatric Urology, Rady Children's Hospital, 7920 Frost Street, Suite 200, San Diego, CA 92123, USA
[b] Department of Pathology, Rady Children's Hospital, 3020 Children's Way, San Diego, CA 92123, USA

ARTICLE INFO

Article history:
Received 16 July 2015
Received in revised form
19 August 2015
Accepted 20 August 2015

Key words:
Lymphangioma
Bladder
Scrotum
Childhood

ABSTRACT

Lymphangioma of the urogenital system is extremely rare and we present two cases treated at our institution over a one year period. The first case is a 3 year-old boy who presented with scrotal swelling and was initially thought to have a complex hydrocele. On surgical exploration, an extratesticular multiloculated cystic mass was discovered and testis-sparing excision of the mass was performed. Pathology revealed lymphangioma. The second case is a 5 year-old male who initially presented with gross hematuria. Ultrasound was unremarkable but cystoscopy revealed varicosities extending from the bladder wall. On transurethral resection, histology showed non-specific benign vascular malformation. Magnetic resonance imaging (MRI) was obtained because of persistent hematuria and showed multiple bladder lesions suggestive of lymphatic malformation. Partial cystectomy was ultimately performed and histology confirmed lymphangioma. To the author's knowledge, this represents the fifth reported case of lymphangioma of bladder.

Lymphangioma is a benign lymphatic malformation, most commonly seen in the neck and axillary regions. Lymphangioma involving the urogenital system is extremely rare. Herein, we describe one case involving the bladder and another case involving the scrotum.

1. Case report 1

A 3 year-old boy initially presented with a 1-year history of painless, progressive swelling in his left scrotum. Physical examination showed bilaterally descended testes and a non-tender, soft, compressible mass in the left hemiscrotum suggestive of hydrocele. Ultrasound of the testes was done and demonstrated fluid around the left testis containing low-level echoes due to debris and multiple septae (Fig. 1). Based on clinico-radiological findings, a provisional diagnosis of complex left hydrocele was suggested. The swelling was initially explored via a left inguinal incision. A multicystic lesion separate from the left testis but densely adhered to the scrotal wall and perineum was identified (Fig. 2). The left testis and spermatic cord were easily separated from the lesion. The left testis was normal in size and appearance. No hydrocele or hernia were identified. Complete excision of the lesion was performed and the overlying scrotal skin spared. Macroscopy showed tan to dark brown to black nodular tissue measuring 4 × 3.8 × 2.7 cm. Sectioning revealed multicystic compartments, some of which contained clear serous fluid, hemorrhagic dark brown material, and clear gelatinous material. Dilated cystic spaces were lined by bland endothelial cells, with small amounts of smooth muscle within the walls of vascular structures (Fig. 3). Immunostaining was positive for CD31 and D2-30, consistent with a diagnosis of lymphangioma. At follow up 1 month later, there was no evidence of recurrence.

2. Case report 2

A 5 year-old boy initially presented with penile pain and blood at the urethral meatus. Physical examination was unremarkable, urinalysis was negative, and a diagnosis of probable meatitis was given. The patient subsequently re-presented three years later at the age of 8 with intermittent gross hematuria. He had no additional complaints. Physical exam was again unremarkable. Urinalysis showed blood but was otherwise negative. Urine culture was negative for bacteria and ultrasonography showed no abnormalities. Cystoscopy under anesthesia was performed, which revealed a lesion with varicosities at the right lateral bladder wall (Fig. 4). Bilateral retrograde pyelograms revealed no upper tract abnormalities. Transurethral resection of the bladder lesion was performed and histology revealed benign vascular malformation. The specimen showed submucosa with varying-sized vascular channels

* Corresponding author. Tel.: +1 858 9668446.
E-mail address: myap@rchsd.org (M. Yap).

http://dx.doi.org/10.1016/j.epsc.2015.08.012

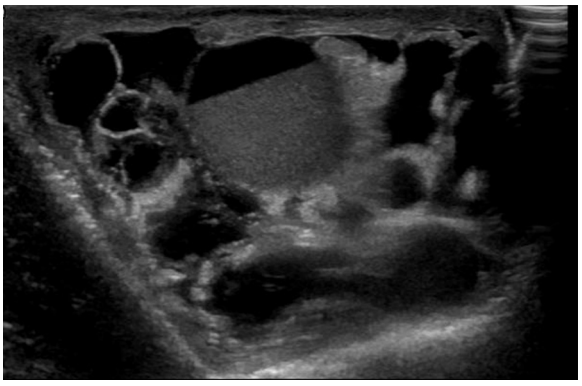

Fig. 1. Scrotal ultrasound showing lymphangioma of scrotum with air-fluid levels.

lined by flat endothelial cells and filled with blood/proteinaceous material. Immunostaining was strongly positive for CD31 and rare for D2-40. Urine cytology was negative. Follow up magnetic resonance imaging (MRI) was performed and revealed multiple nodular and serpiginous areas of T1 and T2 signal abnormality, extensively involving the right lateral wall, left anterolateral wall, dome, and bladder base (Fig. 5). The lesions exhibited inconsistent signal changes but the bulk appeared bright on T1 and T2 sequences and did not enhance. The dominant bladder lesion measured approximately 26 × 9 × 21 mm. On magnetic resonance angiogram (MRA), no arterial or venous contribution, early draining vein, or early arterial enhancement were seen to suggest venous malformation, arteriovenous malformation (AVM), or hemangioma. Surgical treatment was undertaken because of the patient's persistent hematuria. Open partial cystectomy was successfully performed. The lesion had poorly defined borders and consisted of many cysts separated from each other by solid partitions and containing transparent fluid. The lesion was primarily located at the bladder dome and anterior wall but also extended to both lateral walls and the posterior bladder wall. The lesions involved the perivesical space and also protruded full-thickness through the bladder at various locations. Histologic examination revealed that the cystic mass was composed of numerous transmural cavernous vascular lesions, some with rare thickened hyalinized or myxoid walls but the majority lined with a single layer of flattened endothelium. Many of the vascular channels contained lightly eosinophilic lymph, while many others were filled with red cells (Fig. 6). Immunostaining was positive for both CD43 and D2-40. The presence of

Fig. 2. Intraoperative view showing multiloculated cystic mass delivered through scrotal incision.

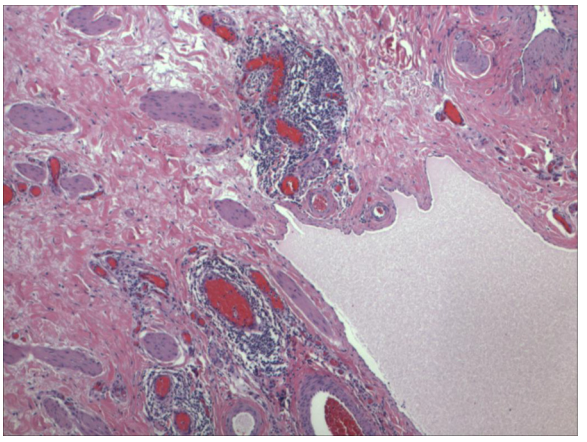

Fig. 3. Scrotal lymphangioma. Large thin-walled vascular space lined by endothelial cells with small amount of smooth muscle and focal collections of lymphocytes in wall.

peripheral blood within some of the lesional vessels raised the diagnostic consideration of a venous component; however, D2-40 labeled the majority of the vessels, supporting the clinical impression of a lymphangioma. The boy had an uneventful post-operative course and was discharged from the hospital 2 days after the procedure. At follow-up 7 months later, patient reported intermittent hematuria that was significantly improved from prior to surgery. A repeat MRI showed residual lesions at the right anterolateral wall decreased from prior to surgery and no new masses. He is being observed for now with plans for possible endoscopic intervention should his hematuria persist.

3. Discussion

Lymphangiomas are benign vascular lesions that usually arise from embryologic disturbances in the development of the lymphatic system [1]. Whereas the majority of these lesions are congenital due to insufficiency or atresia of the efferent lymphatics and venous channels, they can also be acquired after infection, inflammation, or degeneration [2]. Symptoms of lymphangiomas are related to the anatomical location of these lesions, as well as to the extent of involvement of the local anatomical structures. They can be seen in any anatomic region but are more commonly seen in lymphatic-rich areas, such as the neck (75%) and axilla (20%) [3]. Histologically, there are three primary types: capillary, cavernous, and cystic. The cystic form is the most common variety [4]. Lesions are composed of vascular spaces filled with eosinophilic and protein-rich fluid. A single layer of flattened endothelium lines the

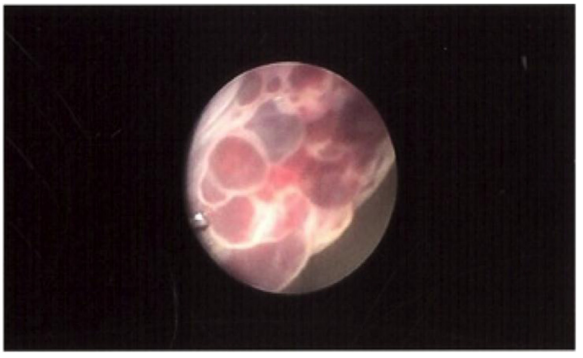

Fig. 4. Cystoscopic view of lymphangioma of the bladder consisting of mucosal varicosities.

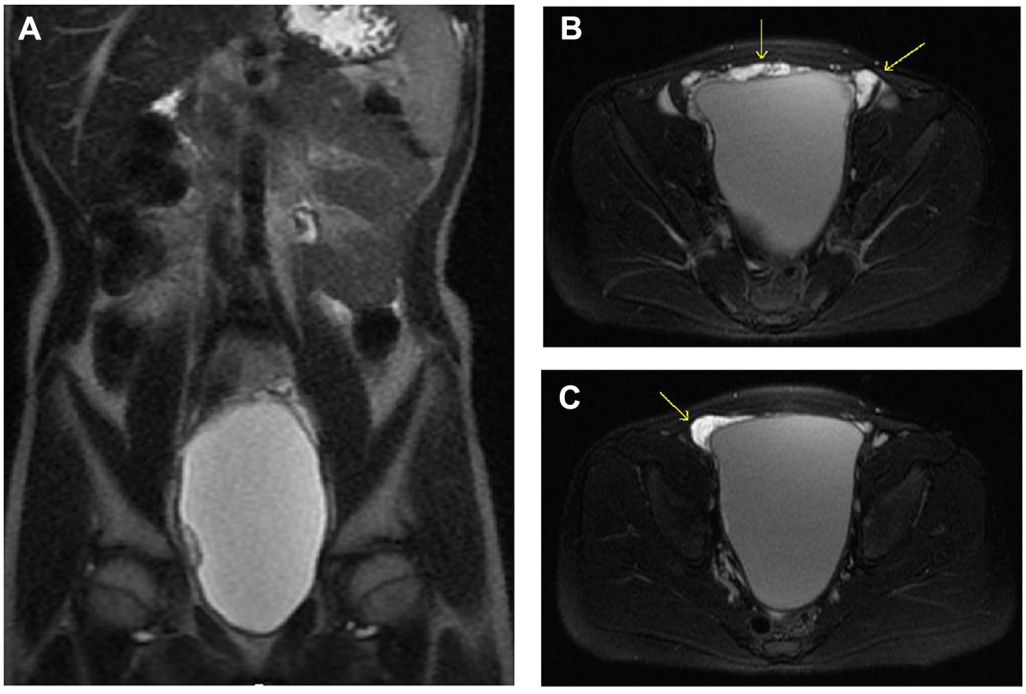

Fig. 5. A) MRI showing lymphangioma of the bladder at right lateral wall. B) MRI showing lymphangioma of the bladder at anterior wall in the midline and left anterolateral wall. C) MRI showing lymphangioma of the bladder at right anterolateral wall.

walls of lymphatic channels. Hemorrhage within the cystic spaces is common and indicates recent trauma or spontaneous intralesional bleeding. The majority of lymphangiomas (90%) develop during the first two years of life and 50% are present at birth [5]. Unlike hemangiomas, lymphangiomas persist throughout life, grow proportionately with the size of the patient, and do not undergo involution [1].

3.1. *Lymphangioma of the scrotum*

The intrascrotal presentation of lymphangiomas is rare at any age. Loberant et al. estimated less than fifty cases reported in the literature as of 2002 [6]. The most typical locations are the scrotal wall and tunics [7], although the testis, epididymis, spermatic cord, and Colles fascia may also be involved [2]. Most present as painless, indolent scrotal masses but can sometimes be associated with acute scrotum due to enlargement or pain following infection or hemorrhage [8,9]. High resolution ultrasonography is highly accurate in delineating the type and extent of lesions in most cases. Ultrasonography reveals a cystic mass with multiple septations and locules filled with low level internal echoes that may be mobile. The echoes may be due to internal hemorrhage or debris. Septations often show low velocity and high resistance vascular flow, characteristic of benign lesions [6]. Computed tomography and magnetic resonance imaging may also be helpful in evaluating for extension into the pelvis or retroperitoneum. As cystic lymphangiomas do not communicate with the lymphatic system, lymphangiography is not recommended [10]. The differential diagnosis of scrotal lymphangioma includes hernia, hydrocele, pyocele, hematocele, spermatocele, varicocele, and primary tumor of the testis [8]. Presence of septal flow differentiates cystic lymphangioma from hydrocele, pyocele, and hematocele; whereas, absence of color flow in the cystic spaces differentiates lymphangioma from varicocele. Despite preoperative imaging, they are often misdiagnosed before surgery [11–13]. Hurwitz et al. reported seven cases of scrotal lymphangioma over a ten-year period, all of which were misdiagnosed prior to surgery [13]. As in our case, the patient was thought to have a complex hydrocele based on clinical and radiologic findings prior

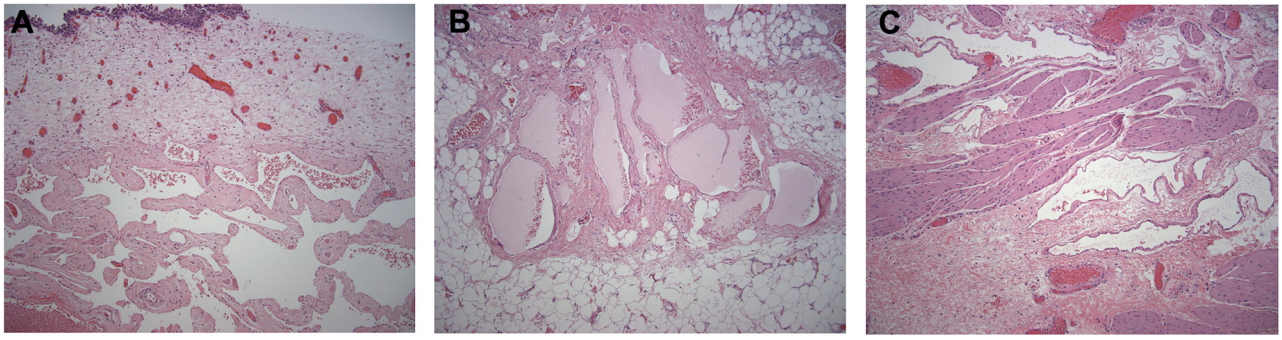

Fig. 6. A) Lymphangioma of the bladder. Submucosal cavernous thin-walled channels. B) Lymphangioma of the bladder. Cavernous thin-walled channels containing lymph and extending into surrounding connective tissue. C) Lymphangioma of the bladder. Cavernous thin-walled channels traversing through mural smooth muscle.

to surgery. Local recurrence is common and complete surgical excision along with overlying skin has been recommended. Other treatment modalities such as injection of sclerosants, extensive fulguration, and cryotherapy have been reported but without much success.

3.2. Lymphangioma of the bladder

Lymphangioma only rarely occurs in the urinary tract. About 40 cases have been described involving the kidney and 2 cases have been reported involving the urethra [14–16]. Only 4 cases of lymphangioma of the bladder have been reported worldwide since 1983. Bolkier et al. reported the first case in a boy who presented with macroscopic painless hematuria and was treated with partial cystectomy [17]. Wyler et al. reported the second case in a 49 year-old male presenting with irritative voiding symptoms. Cystoscopy showed a non-papillary lesion located between the two ureteral orifices. The patient was managed with transurethral resection and histology revealed lymphangioma of the bladder. Three months after the procedure, the irritative voiding symptoms had improved and there was no evidence of residual tumor on cystoscopy [18]. Niu et al. reported on two additional cases of bladder lymphangioma in children. The first was an 8 year-old girl who presented with terminal hematuria accompanied and intermittent fever. Imaging revealed a sharply defined 0.5-cm mass in the right lateral bladder wall. On cystoscopy, the tumor was red and found to be bulging into the bladder. A partial cystectomy was performed and histology revealed lymphangioma involving the full-thickness of the bladder. She had no evidence of recurrence at 3 years of follow up [19]. The other case reported by Niu et al. was a 5 year-old boy who presented with abdominal pain and palpable pelvic mass. Computed tomography and magnetic resonance imaging revealed a 12-cm cystic lesion anterior to the rectum, occupying the pelvic cavity, and displacing the bladder downward and laterally. Mass partial cystectomy was performed and histology revealed lymphangioma. The mass was located extraperitoneally and was firmly attached to the superior and posterior walls of the bladder. It penetrated the detrusor muscle but did not protrude into the bladder cavity. The boy was recurrence-free at 3 months [20]. While not specifically described for treatment of bladder lymphangiomas, endoscopic laser ablation may be an option based on reports of successful control of bladder hemangiomas, mainly using the neodymium:yttrium aluminum garnet (Nd:YAG) laser. It is important to know that when laser ablation has been used for bladder hemangiomas, treatments were often carried out several occasions [21–23].

4. Conclusion

In summary, although the scrotum is a very rare site for lymphangioma, it should be considered in the list of differential diagnosis of multiloculated, benign-appearing scrotal lesions. Although ultrasonography can be useful in establishing a preoperative diagnosis, misdiagnosis is relatively common. Awareness of these lesions is integral in making a correct diagnosis, to avoid mismanagement and reduce recurrences.

Lymphangioma of the bladder is also a benign lesion and does not regress. Its symptoms are generally related to infection and hemorrhage, as well as to the size of the malformation and perturbation of adjacent tissues altering structure and function. Open resection is ideal for the disease; however, because the lesion is benign, an extensive operation is not warranted. Endoscopic laser ablation may be an option when partial cystectomy is precluded but may require multiple procedures for initial control and repeat procedures for recurrences. Preoperative diagnosis may be difficult because of the rarity of this lesion and it's non-specific presentation clinically and radiologically. Ultrasound has been suggested as a useful tool for follow up in children and prognosis is generally good after resection.

Conflict of interest

None.

References

[1] Elleru RG, Balakrishnan K, Padua HM. Lymphatic malformations: diagnosis and management. Semin Pedaitr Surg 2014;23:178–85.
[2] Koshy A, Tandon RK, Kapur BM, Rao KV, Joshi K. Retroperitoneal lymphangioma: a case report with review of the literature. Am J Gastroenterol 1978;69:485–90.
[3] De Perrot M, Rostan O, Morel P, Le Coultre C. Abdominal lymphangioma in adults and children. Br J Surg 1998;85:395–7.
[4] Cutillo DP, Swayne LC, Cucco J, Dougan H. CT and MR imaging in cystic abdominal lymphangiomatosis. J Comput Assist Tomogr 1989;13:534–6.
[5] Mulcahy JJ, SChileru G, Donmezer M, Bhathena D. Lymphangioma of the scrotum. Urology 1979;14:64–5.
[6] Loberant N, Chernihovski A, Goldfeld M. Role of Doppler sonography in the diagnosis of cystic lymphangioma of the scrotum. J Clin Ultrasound 2002;30:384–7.
[7] Millan RW, Donald BR, Alpern HD. Scrotal lymphangioma. Urology 1984;33:79–80.
[8] Wiedman ED, Cendron M, Schned AR, Harris RD. Scrotal lymphangioma: an uncommon cause for a scrotal mass. J Ultrasound Med 2002;21(6):669–72.
[9] Hacker A, Hatzinger M, Grobholz R, Alken P, Hoang-Bohm J. Scrotal lymphangioma: a rare cause of acute scrotal pain in childhood. Aktuelle Urol 2006;37:445–8.
[10] Chung SE, Frush DP, Fordham LA. Sonographic appearance of extratesticular fluid and fluid containing scrotal masses in infants and children: clues to diagnosis. AJR Am J Roentgenol 1999;173:741–5.
[11] Ratan SK, Rattan KN, Sehgal T, Maggu S. Spermatic cord lymphangioma in a 7 year old child masquerading as a hydrocele. Br J Urol Int 2003;10:4046.
[12] Parakh A, Dubey AP, Garg A, Khurana N, Aggarwal SK. Cystic lymphangiomatous hamartoma masquerading as massive ascites. Indian J Pediatr 2009;76:753–4.
[13] Hurwitz RS, Shapiro E, Hulbert WC. Scrotal cystic lymphangioma: the misdiagnosed scrotal mass. J Urol 1997;158:1182–5.
[14] Honma Y, Takagi Y, Shigyo M, Sunaoshi K, Wakabayashi J, Harada O, et al. Lymphangioma of the kidney. Int J Urol 2002;9:178–82.
[15] Jadeja NA, Yadav SP. Lymphangioma of the urethra. Urol Int 1994;53:104–6.
[16] Tahtali N, Yazicioglu A, Dalva I, Sargin S, Cetin S. An unusual cause of urethral stricture: urethral lymphangioma. Eur Urol 1994;26:347–8.
[17] Bolkier M, Ginesin Y, Lichtig C, Levin DR. Lymphangioma of bladder. J Urol 1983;129:1049–50.
[18] Wyler SF, Bachmann a, Singer G, Gasser TC, Sulser T. First case of lymphangioma of the bladder in an adult. Urol Int 2004;73(4):374–5.
[19] Niu ZB, Yang Y, Hou Y, Chen H, Liu X, Wang CL. Lymphangioma of bladder. Urology 2010;76(4):955–7.
[20] Niu ZB, Ying H, Sun RG, et al. Cystic lymphatic malformation of bladder presenting as a pelvic mass. J Pediatr Surg 2011;46:1284–7.
[21] Takemoto J, Yamazaki Y, Sakai K. A case of large bladder hemangioma successfully treated with endoscopic yttrium aluminum garnet laser irradiation. Int J Urol 2011;18:854–6.
[22] Vicente J, Salvador J. Neodymium:YAG laser treatment of bladder hemangiomas. Urology 1990;36:305–8.
[23] Smith Jr JA, Dixon JA. Neodymium:YAG laser irradiation of bladder hemangioma. Urology 1984;24:134–6.